

Title: Marfan Syndrome *GeneReview* – Management: Animal Models and Losartan

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Note: The following information is provided by the author and has not been reviewed by *GeneReviews* staff.

## **Animal Models – Use of Losartan**

Studies in animal models of Marfan syndrome demonstrated excessive activation of and signaling by the growth factor TGF $\beta$ . Systemic administration of TGF $\beta$  antagonists can attenuate or prevent many disease manifestations in fibrillin-1-deficient mice including emphysema, skeletal muscle myopathy, myxomatous valve disease, and aortic aneurysm. Losartan, an angiotensin II type 1 receptor blocker, can also decrease TGF $\beta$  signaling. Losartan has shown the ability to halt abnormal aortic root growth in mouse models of Marfan syndrome [Habashi et al 2006]. This effect associates with both a reduction in hemodynamic stress and antagonism of TGF $\beta$  signaling in the vessel wall.

## **References**

Habashi JP, Judge DP, Holm TM, Cohn RD, Loeys BL, Cooper TK, Myers L, Klein EC, Liu G, Calvi C, Podowski M, Neptune ER, Halushka MK, Bedja D, Gabrielson K, Rifkin DB, Carta L, Ramirez F, Huso DL, Dietz HC. Losartan, an AT1 antagonist, prevents aortic aneurysm in a mouse model in Marfan syndrome. *Science* 2006;312:117-21.